

Idiopathic intracranial hypertension; incidence, presenting features and outcome in Northern Ireland (1991-1995)

J J Craig, D A Mulholland, J M Gibson

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SUMMARY

Objectives: to determine the age and sex specific incidence rates, presenting features, and visual outcome of idiopathic intracranial hypertension in Northern Ireland.

Methods: A case-note review of all patients with idiopathic intracranial hypertension, diagnosed at the Royal Victoria Hospital, Belfast between 1991 and 1995.

Results: Forty-two patients were identified corresponding to an average annual incidence rate per 100,000 persons of 0.5 for the total and 0.9 for the female population. The commonest presenting symptoms were headache (84%), transient visual obscurations (61%) and sustained visual loss (34%). Impaired Snellen visual acuity and visual field loss were documented in 21% and 62% of patients respectively at presentation, and in 24% and 39% at last follow-up. One patient suffered deterioration in visual functioning sufficient to interfere with normal daily activities. **Conclusions:** The age and sex specific incidence rates of IIH in Northern Ireland are lower than have been reported in previous population-based series. Disabling visual loss occurs in a small number of patients despite all interventions.

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is a condition of unknown aetiology characterized by symptoms and signs of increased intracranial pressure in the absence of an intracranial lesion or hydrocephalus. All neuroscience units manage IIH, yet information on epidemiology is very limited and is based on studies that have involved either small numbers of patients or short periods of follow-up.¹⁻³ Figures for European populations are unknown and some reports have suggested that IIH is less common in Europe.^{4,5}

Visual loss is the most important complication of IIH. Previous studies have suggested that 96% of patients have visual abnormalities at some time,⁶ and up to 10% develop bilateral blindness.⁷ There are no controlled trials of treatments in this condition, and despite all therapeutic interventions some patients develop progressive blindness.

The aims of our study were: (1) to determine the population-based incidence rates of IIH in Northern Ireland, over a five year period (1991-1995), (2) to provide clinical details of cases of IIH diagnosed in our locality over this period,

and (3) to determine the frequency of visual complications in our patients.

CASES AND METHODS

Cases were identified by a computer search of the medical records for all patients treated at the Royal Victoria Hospital (RVH), Belfast between the 1st of January 1991 and 31st of December 1995 for whom the following diagnoses were made; pseudotumour cerebri, benign intracranial hypertension or IIH. The RVH is a university teaching hospital, and houses the only departments of neurology and neurosurgery and the main department of ophthalmology that serve Northern

Department of Neurology, Royal Victoria Hospital, Grosvenor Road, Belfast.

John J Craig, BSc(Hons), MRCP, Specialist Registrar.

J Mark Gibson, MD, FRCP, Consultant.

Department of Ophthalmology, Royal Victoria Hospital, Grosvenor Road, Belfast.

David A Mulholland, MRCP, FRCOphth, Specialist Registrar.

Correspondence to Dr Craig.

Ireland (Population 1.64 million). All patients with IIH would be expected to be seen in this hospital at some stage. Inclusion in the study was based on the Modified Dandy Criteria for IIH (Table 1). To be considered, the diagnosis of IIH should have been made between 1st January 1991 and the 31st of December 1995 and residency in Northern Ireland had to be established at least 1 year before diagnosis.

TABLE I

Modified Dandy Criteria for the diagnosis of idiopathic intracranial hypertension.⁸

1. Signs and symptoms of increased intracranial pressure.
2. Awake and alert patient,
3. No abnormal neurological findings except papilloedema or a sixth nerve palsy.
4. Normal CT/MRI except for empty sella syndrome or small ventricles.
5. Documented increased CSF opening pressure (>200 mm of water in non-obese and >250 mm of water in obese patient), with normal CSF composition.
6. No other known cause of raised intracranial pressure.

Age and sex specific incidence rates were calculated by dividing the number of incident cases by the proper denominator, gained from census data for Northern Ireland 1993. Data were independently collected from the case-notes by two of the authors (JC + DM). The follow-up period was to the 31st May 1996. Variables collected were symptoms, and examination findings, recorded at presentation and on each visit.

RESULTS

Forty-two cases, with a mean age of 29 years at diagnosis, were identified in the five-year study period. Average annual incidence rates per 100,000 persons for IIH by age and sex are shown in table II. The average annual incidence rates per 100,000 were 0.6 for the total and 0.9 for the female population. The female-to-male ratio of incidence rates was 5.7:1.0.

Symptoms (Table III) Duration of symptoms prior to diagnosis varied from one to 208 weeks. Headache was the presenting symptom in 32 cases and occurred daily in almost two-thirds. Transient visual obscurations (TVOs) were reported by five of the six cases that did not have headache. Half reported weight gain in the year before diagnosis.

Eleven (26%) patients had magnetic resonance (MR) imaging of brain with MR venography. These were reported as normal for all cases.

Table II

Age and sex specific average annual incidence rates of idiopathic intracranial hypertension per 100,000 population in Northern Ireland, 1991 to 1995.

Age Group, years	Females		Males		Total	
	No.	Rate	No.	Rate	No.	Rate
0-14	2	0.20	1	0.10	3	0.10
15-24	12	1.98	0	0.00	12	0.96
24-35	10	1.56	2	0.32	12	0.93
35-44	7	1.33	1	0.19	8	0.77
45+	5	0.35	2	0.16	7	0.26
All age groups	36	0.86	6	0.15	42	0.51

TABLE III

Presenting symptoms

Symptom	%
Headache	84
Transient visual obscurations	61
Sustained visual loss	34
Scintillations	18
Diplopia	11
Intracranial noises	8
Retrobulbar pain	5
Deafness	3
Weight gain	50

Ophthalmological Examination All patients had optic disc swelling at presentation (Figure 1). In eleven cases it was mild and in 21 moderate to severe. Corrected Snellen visual acuity at presentation was worse than 6/9 in at least one eye in eight (21%) patients. An abnormality other than enlargement of the blind spot was recorded in 18 (62%) of the 29 who had perimetric assessment of visual fields performed at presentation. The commonest field abnormalities noted were generalized constriction and infero-nasal field defects (Figure 2).

Sixteen (42%) cases had persistent disc swelling (ten mild, six chronic) at last follow-up. Corrected Snellen visual acuity was worse than 6/9 in at least one eye in nine (24%) cases. Four of these nine were cases whose visual acuity had been normal at presentation. None had deterioration of visual acuity sufficient to interfere with normal daily activities. Seven (39%) of the 18 cases who had visual field loss documented at presentation had complete resolution of their defects. One case had a worsening of field loss with marked field constriction to within 20 degrees of fixation, which interfered with activities of daily living. The rest maintained persistent mild defects that did not interfere with visual functioning.

Patients were treated with acetazolamide or diuretics initially. Analgesic preparations taken on a regular basis by almost three-quarters of the patients were never effective at relieving headache. Over half also had repeated lumbar punctures. Weight loss was documented in eight

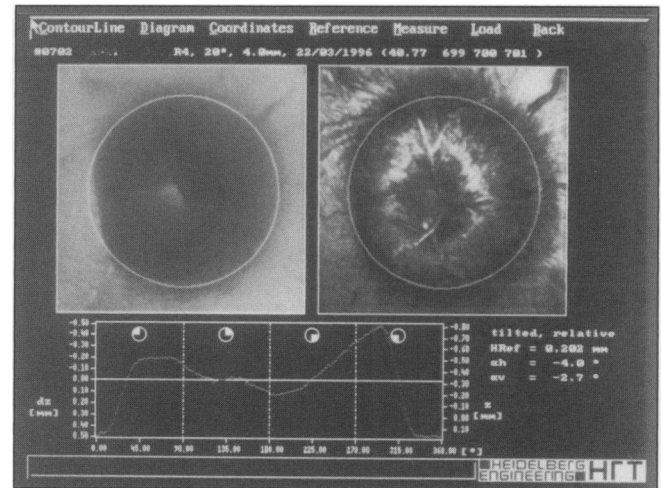


Fig 1. Confocal laser tomogram of right eye of patient showing significant disc swelling.

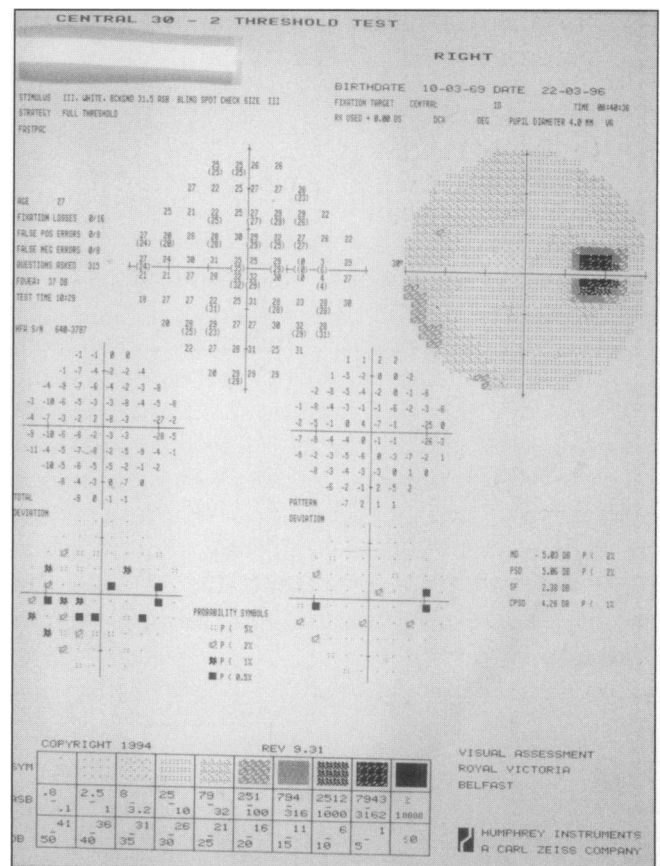


Fig 2. Humphrey visual field of central 30° of vision of right eye of patient showing slight enlargement of the blind spot and early infero-nasal field loss, which is often reversible.

of the 21 patients reporting significant weight gain prior to symptom onset. This was sustained in four, with three having sustained symptomatic improvement and reduced CSF opening pressure. Seven cases (18%) had a theco-peritoneal shunt inserted. Optic nerve sheath fenestration was not performed in any of the cases. Symptomatic improvement occurred in all patients who had a shunting procedure, which was maintained for a mean of 16 months. All patients who had surgical management developed post-operative problems, including a return of symptoms of raised intracranial pressure in six patients; one of these patients also had a shunt infection. The other patient developed chronic headache due to persistently low cerebrospinal fluid pressure. Further surgery, amounting to 15 additional procedures, was necessary in four of these six patients.

DISCUSSION

The average annual incidence rates of symptomatic IIH in Northern Ireland per 100,000 persons at 0.5 for the total population and 0.9 for females are lower than those reported in the three previously conducted population based studies evaluating the incidence of this condition.¹⁻³ In these studies the incidences per 100,000 ranged from 0.9 to 1.7 for the total population and 1.6 to 3.6 for females. Whether our results are due to incomplete ascertainment or that the incidence of IIH is lower in our locality, due to such factors as the higher frequency of obesity in the populations previously studied, is unknown. Patients with papilloedema and headache without tumour will be referred to our single neuroscience centre; diagnosis is precise and our population denominator is stable. Thus we feel that the observed rates are representative of the true incidence rates of symptomatic IIH in Northern Ireland.

Headache was less often the presenting symptom in our cases. Most of those who did not have headache had TVOs. TVOs without headache have been documented in IIH⁹ but never as frequently as in our series. This highlights the importance of establishing whether papilloedema and raised intracranial pressure are present in patients with TVOs.

Visual acuity and visual fields were impaired in a substantial number of patients at presentation reflecting the delay between symptom onset and diagnosis. Visual loss severe enough to interfere

with activities of daily living was however uncommon at any stage in our cases. In addition the abnormalities found on testing visual functioning often improved. Our results are in keeping with those of the population-based series from Rochester, Minnesota³ where nine patients identified over a 15 year period from a population of 70,000 were followed up for a median period of 2.7 years. Out of 18 eyes only three developed visual impairment. This was mild in all cases and did not interfere with everyday visual functioning. Such findings are at odds with the commonly held belief that disabling visual loss is a frequent result of IIH,⁶ and might be explained by the relatively short follow-up period of our cases and those from Rochester. However, some of the difference could also be accounted for by selection bias in non-population-based studies with more severely affected cases being studied.

Efficacy is claimed for medical treatments used in IIH.^{6, 10-14} There are however no controlled trials of any of them and patients are therefore managed according to personal or local preferences. This is also the case for the surgical procedures used. There is no doubt that surgical intervention has saved vision in some patients with IIH¹⁵⁻¹⁷ but the effectiveness of either shunting procedures or optic nerve sheath fenestration have never been studied in a controlled way, and it is unclear when surgical intervention should be undertaken. Both types of procedure are also not infrequently associated with post-operative complications;¹⁸⁻²² some patients may therefore be having an unnecessary intervention with attendant significant morbidity.

In conclusion, we found that the age and sex specific incidence rates of IIH are lower in Northern Ireland than in previous population-based studies and that visual loss sufficient to interfere with normal activities was uncommon. In keeping with the results from Rochester, Minnesota, this raises the possibility that the prognosis for vision in IIH is better than previously suggested from the results of non-population based series.

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